

Rheumatic fever

Rheumatic Fever

- RF is generally classified as connective tissue (CT.) or collagen vascular disease, and can be defined as: - an inflammatory reaction that involve heart, joints CNS & subcutaneous tissue.
- The major importance of RF is its ability to cause fibrosis of the heart valves leading to crippling hemodynamics of chronic heart disease.

Predisposing factors

1. **Age:-** risk of RF occurs commonly between the age 5-15 y.
2. **Sex:** - both sexes are affected equally but chorea is common in females.
3. **Familial predisposition:** -RF may run in families due to common environmental factors as overcrowding & substandard housing which predisposes to streptococcal pharyngeal infection.
4. **Season:** -it is common in colder & wetter months
5. **recurrent strept. Infection.**

Pathogenesis

- *As many as 3% of untreated acute Streptococcal sore throat may be followed by RF*
- *1/3 of acute RF are well documented to follow mild asymptomatic pharyngitis*
- *RF does follow strept skin infection*
- *Strains of group A strept. (GAS) that have been associated 'with outbreaks of RF have virulent properties such as rich content of type specific surface M protein (which share a long terminal antigenic domain & contain epitopes that are shared with human heart tissue particularly sacrolemmal membrane proteins & cardiac myosin) & large hyaluronidase capsule which is highly resistant "to phagocytosis & induce vigorous immune responses.*
- *several strept. 'Antigens (ag) have demonstrated cross reactivity with cardiac and other tissue ...antibodies (ab) formed against these Ag therefore react with these tissues resulting in their damage*

Generally the pathogenesis of RF depends on the etiological agent the host.. see the following table:-

Group A strept.

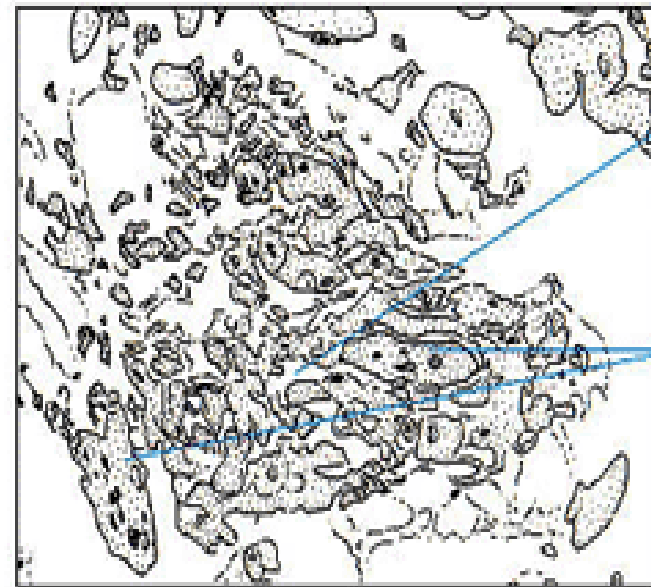
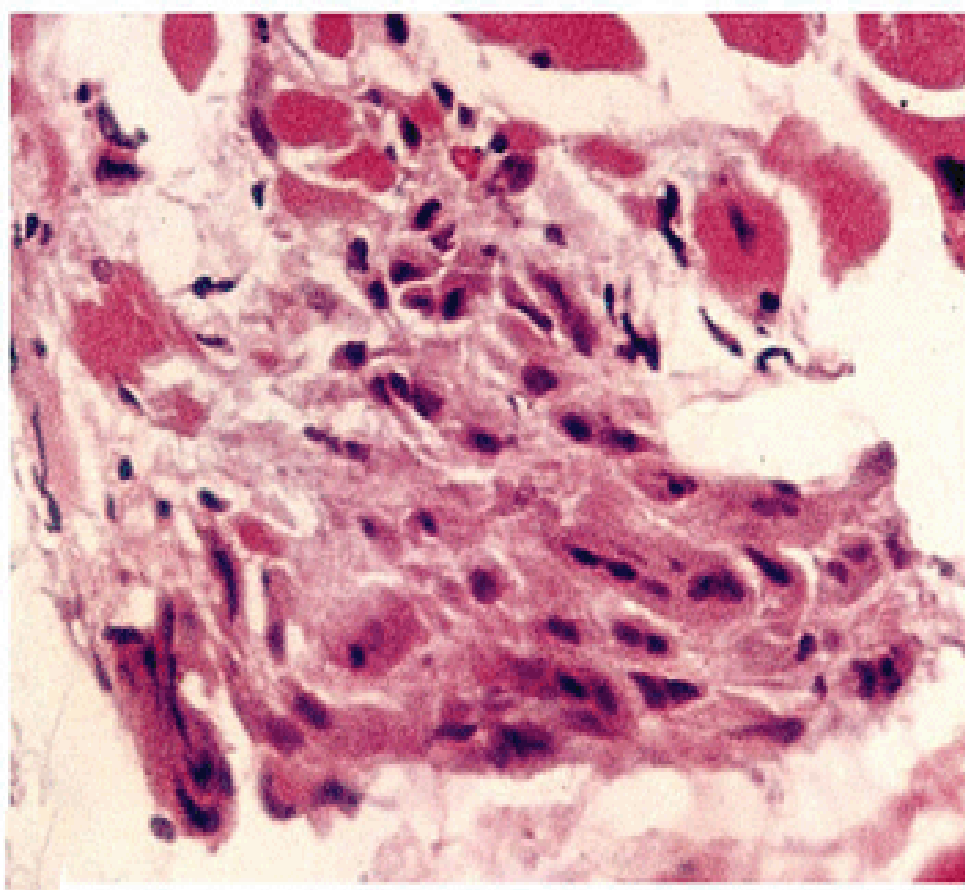
- ▶ **tonsillopharyngeal infection not other site**
- ⌚ **intensity of the infection**
 - 1.brisk ab response**
 - 2. persistence of the microorganism**
- **rheumatogenic strain. M types 1.3.5.6.14.18..19.27.&29**
- **Heavily encapsulated forming mucoid colonies.**
- **resist phagocytosis.**
- **do not produce opacity factor**

Susceptible host

- **genetic predisposition**
 - **presence of specific B cell alloantigene**
 - **high incidence of class II HLA antigen**

Pathology

- *The acute phase of RF characterized by exudative & proliferative inflammatory reactions involving C.T or collagen tissue mostly in the heart, brain, joints, & subcutaneous tissue.*
- *Generalized vasculitis affecting small blood vessels is commonly noted but unlike vasculitis of other CT disease thrombotic lesions are not seen .*
- *Aschoff nodules (autopsies finding) :-*
 - 1.pathognomonic of Rh. Carditis*
 - 2.Found in the myocardium.*
 - 3.Persist for years after acute attack.*
- *Edema & cellular infiltration of the valvular tissue & chordae tendinae followed by hyaline degeneration of the affected valve lead to the formation of verrucae at it's edge which prevent total closure of the valves. Finally fibrosis and calcification of the valve occur & end in valvular stenosis.*



Fibrinoid necrosis

*Multinucleated
Aschoff giant cells*

Aschoff nodules (autopsies finding)

Diagnosis

- *Duckett Jones formulated his criteria for the diagnosis of RF through*
 - ***2 major criteria*
 - *** 1 major + 2 minor criteria plus evidence of previous GAS infection*

| Major Criteria | Minor Criteria | <i>Evidence of preceded GAS infection</i> |
|-----------------------------|---------------------|---|
| <i>Carditis</i> | <i>Fever</i> | <i>Positive throat culture</i> |
| <i>Arthritis</i> | <i>Arthralgia</i> | <i>Positive rapid strept. Ag test</i> |
| <i>Sydenham chorea</i> | <i>CRP</i> | <i>Increased strept. Ab titer</i> |
| <i>Erythema marginatum</i> | <i>High ESR</i> | <i>Previous scarlet fever</i> |
| <i>Subcutaneous nodules</i> | <i>Prolonged PR</i> | |

Carditis

- *The most specific manifestation of RF noted in 50% of pt.*
- *It is pancarditis affecting endo, myo, and pericardium*
- *Valvulitis (endocarditis) involving mitral & aortic valves & the chordae tendinae of the mitral valve is the most characteristic component of Rh carditis AR is less common and associated with MR... pulmonary & tricuspid valves are rarely involved.*
- *Myocarditis present in the following:*
 - tachycardia disproportionate to the degree of fever
 - gallop rhythm
 - rapid cardiac enlargement
 - HF

■ *Pericarditis* which could be :-

1.dry carditis.

2. Pericardial effusion.

3.Never constrictive pericarditis.

■ *Myocarditis or pericarditis in the absence of valvulitis is **not** likely to be due to RF.*

Arthritis

- *The most common but less specific manifestation*
- *Asymmetrical, affecting large joints (knee ,elbow, wrist)*
- *There is swelling, redness, hotness, pain, and limitation of movement &tenderness on touch.*
- *Migratory, **not** result in permanent deformity*
- *If untreated it last 2-3 weeks*
- *Striking feature of Rh arthritis is its response to salicylate.*
- *Indeed if a pt, Does not improve substantially after 48 h. of adequate salicylate treatment the diagnosis of RP should be in doubt*
- *Some pt may develop arthritis & other manifestation following acute strept. Pharyngitis that do not fulfill the jones criteria for the diagnosis of RP. this syndrom is called poststreptococcal reactive arthritis (PSRA) which does not respond dramatically to salicylate*
- *Some pt. With PSRA may have silent or delayed onset carditis, therefor these pt, Should be observed carefully .for several months for the subsequent development of Carditis.*

Chorea

- *Called sydenham chorea or saint vitus dance,*
- *Seen In 20%. Of RF patients*
- *It is due to an inflammatory process involving basal ganglia & caudate nuclei.*
- *It appears in 3 months or longer after strept infection while. carditis & arthritis appear within 3 weeks.*
- *Chorea characterized by :*
 - *Spontaneous, semipurposeful & non-repetitive movements.*
 - *Occur at rest increased by emotion & disappears with sleep*
 - *Associated with hypotonia & emotional lability.*
 - *knee jerk may be pendular .*
 - *speech & handwriting deteriorate*
 - *Resolve in 1-2 weeks even without treatment.*

Erythema marginatum

- *Occur in <5%.*
- *Lesions vary greatly in size and Present on the trunk & proximal extremities, but not in the face.*
- *It is an envascent erythema, macular, non pruritic rash with pale centers & rounded or serpiginous margin.*
- *It is transient & may be brought out by heat application.*



Subcutaneous nodules

- *Occur in 3%.*
- *Mostly seen in pt. With carditis*
- *They are painless, firm, nodules that measure 0.5- 2cm over the extensor surface over the joints (elbow, knee & wrists) in occipital area of the scalp or over spinous process.*

Clinical findings

- *Fever & arthralgia are non-specific, common findings in pt. With acute RF.*
- *Their diagnostic value is limited.*
- *They are used to support the diagnosis of RF when only a single major manifestation is present.*
- *Epistaxis & abdominal pain may also occur but are not include as minor criteria.*

Lab. findings

- *Mild normochromic anemia & leukocytosis*
- *CRP is always elevated during acute phase but usually normal in pt. With chorea.*
- *ESR is useful in following the course of the disease but it could be falsely elevated if pt had anemia , and falsely low if pt had already HF. (unlike ESR CRP is not affected by anemia or HF)*

ANTECEDENT GAS INFECTION:-

- *It is important to demonstrate GAS in the pharynx or the presence of elevated strept ab titer*
- *several rapid GAS ag detection tests are available but although they have high degree specificity, they have low sensitivity .*
- *Increased GAS ab titer produce more reliable evidence of recent injection. The most commonly used is ASOT (significant if >250 Todd unit, anti-DNase -B is also available.*

- *Chest X-ray could be normal, cardiomegaly, pericarditis, pulmo. Edema can also be seen.*
- *ECG shows :- prolonged PR, tachycardia, ST-T change, atrioventricular block.*

Treatment

GENERAL

- *Hospitalization*
- *Bed rest until ESR & CRP return to normal.. In pt. With carditis rest should continued for 2-6 weeks after these parameters return to normal.*
- *10 days course of oral penicillin should be given OR a single dose of benzathin penicillin. (Erythromycin or cephalosporins can be used in pt. who are sensitive to penicillin.)*
- *Antifailure therapy if heart failure is present*
- *Valve replacement may be needed in severe valvular damage.*

PREVENTIVE therapy

- **1.PRIMARY prevention** :- by early detection & treatment of strept infection by Benzathine penicillin single IM in}.
600,000 u for pt <27kg or 1.200.000 u for pt. >27 kg
- **2.SECONDARY prevention** :- by long acting penicillin
1.200.000 u / 3 weeks For how long acting penicillin should be continued?
- See the following table: -

| Category | duration |
|--|---|
| <i>RF with carditis & residual valvular disease</i> | <i>At least 10 y. after last episode & at least until age 40... sometimes longlife.</i> |
| <i>RF with carditis but no residual valvular disease</i> | <i>10 y. or until adult hood</i> |
| <i>RF with out carditis</i> | <i>5 y. or until age 21 y. whichever longer.</i> |

ANTIRHEUMATIC therapy

- *Salyicylate (aspirin) 100 mg/kg/day 4 times daily (max. dose is 8 g /day) to be continued until satisfactory clinical response occur usually dramatic response occur within 48 hours, if not the diagnosis of RF should be re-evaluated) the dose should then be decrease it 2/3 and continue until ESR & CRP normalized.*
- *Corticosteroid is preferable for pt. With carditis in The form of prednisolone 1-2 mg/kg 4 times daily 4 weeks then tapering the dose with addition of aspirin to prevent post steroid rebounds*
- *For RF chorea:- diazepam or hallopridol*

Follow up

- *Carditis most frequently occurs within 2 weeks of arthritis.*
- *Chronic RHD is common in pt. Who have carditis during the initial attack.*
- *Echo. Is valuable in assessing valve problems if it remains normal, follow up can be discontinued 10 y after the initial attack.*